

Neuropathic bladder

Rare

Girls > boys

Boys with more upper tract complications

CISC mainstay of treatment but only ~20% dry

Aetiology

Congenital

Myelomeningocele

Spinal bifida occulta

Sacral agenesis

Acquired

Trauma

Infarction

Tumour

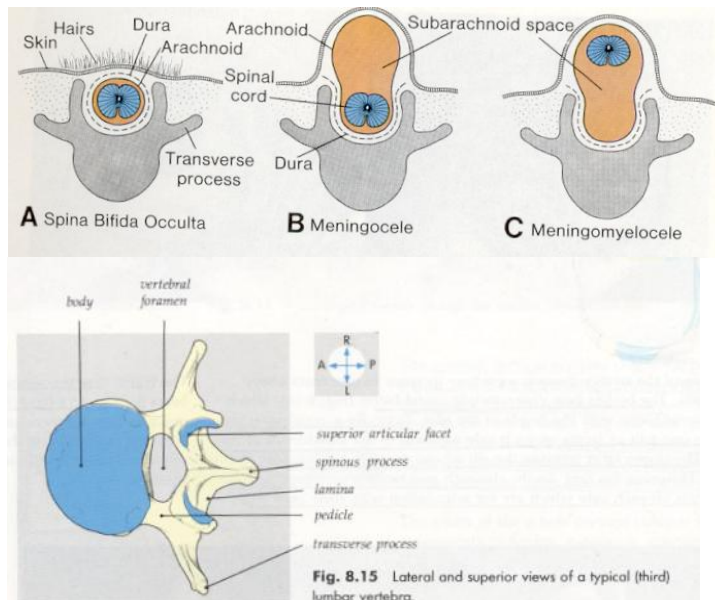
Transverse myelitis

Myelomeningocele (subtype of spinal bifida) represents commonest cause of congenital neuropathic bladder

Failed fusion of dorsal vertebrae (lamina)

Covered by skin - spina bifida occulta. More than one vertebrae causes bulging of meninges – meningocele. Very large sacs contain cord – myelomeningocele.

Conus medullaris invariably involved – 95% have bladder/bowel dysfunction although 30% retain sacral cord reflexes



Common associated findings

Hydrocephalus

Caudal displacement of medulla oblongata (Arnold Chiari malformation)

Kyphoscoliosis

Renal agenesis

Spina bifida occulta

Spinal dysraphism covered by skin, often with a hairy patch or atypical presacral dimple. Atypical presacral dimple defined as offcentre, > 2.5cm from anal verge and > 0.5cm deep. 40% of patients with atypical presacral dimple will have underlying spinal dysraphism. Combination of hairs/atypical dimple a/w highest risk of dysraphism. Diagnosis:

- If < 6 months old – spinal USS (ossification of spine at 6 months)
- If > 6 months old – spinal MRI

Classification of NLUTD in myelomeningocele

Suprasacral (discoordinated voiding)

- Retained sacral conus reflexes (anocutaneous (S5); bulbocavernosus (S3,4))
- Neurogenic detrusor overactivity
- DSD
- No sphincter paralysis

Sacral (acontractile bladder)

- Absent sacral conus reflexes
- Often poorly compliant bladder
- High pressure (retainers) or low pressure (wetters) dependent on intrinsic EUS tone (static sphincteric obstruction)
- NB. Paralysis of EUS means that intrinsic tone may be overcome by external pressure – ‘expressible bladder’
- pathognomonic for neuropathic bladder (Crede’s manoeuvre)

Management neuropathic bladder

Principles

- #1 Preserve renal function
- #2 Provide continence (3 features)
 - (i) adequate storage
 - (ii) complete emptying*
 - (iii) voiding at will*

* CISC in all patients if possible (but only 20% continent)

Additional measures

A. Detrusor hyperreflexia & detrusor non-compliance

- Anticholinergics
- Augmentation cystoplasty
 - Enterocystoplasty
 - Ureterocystoplasty (only grossly dilated ureters)
 - Detrusor myomectomy
 - Detrusor myomectomy + demucosalised enterocystoplasty

B. Sphincteric incompetence

- Medication (α-adrenergic agonists)
- Periurethral bulking agents
- Bladder neck suspension
- Bladder neck slings
- Pippe-Salle (aka Kropp onlay procedure – girls only)
- AUS

Bladder neck closure

c. Other surgical procedures

Mitrofanoff

Appendix, fallopian tube, Monti

Vesicostomy

Urethral sphincterotomy

Ileal conduit

MACE

Exstrophy-epispadias complex

Failure of cloacal membrane to retract caudally prevents medial migration of mesenchyme = rotational abnormality of lower abdomen and pelvis

In order of severity cloacal exstrophy, bladder exstrophy and epispadias

1. Bladder exstrophy

Most common (1:50,000)

Boys > girls (3:1)

Increased risk 500x in offspring of adults with exstrophy

Usually detected by fetal ultrasonography

Features

Low-lying umbilicus

Exposed bladder plate

Pubic symphysis diastasis

Divarication of recti

Inguinal hernia (80% boys; 10% girls)

Anterior ectopia of vagina and anus

Epispadias always present

a/w VUR and inguinal hernia but not other congenital anomalies
(cf. cloacal exstrophy)

Management

Traditionally 3-stage surgical repair:

(i) Primary bladder closure

neonatal period, typically with pelvic osteotomy

no attempt to close bladder neck – all incontinent

(ii) Epispadias repair (see below)

1-2 years

(iii) Bladder neck closure

> 5 years

Bilateral ureteric reimplantation and tubularisation of trigone

Augmentation for those with low bladder capacity

Urethral sensation intact – ISC often problematic
requiring mitrofanoff +/- BN closure

Recently a number of centres (Erlangen perhaps most famous)
have advocated a single-stage repair at ~ 10 weeks

Outcome

70% acceptable continence after staged repair (NB. Single most important factor predicting continence is bladder capacity)

50% upper tract dilatation

Reasonable sexual function in males but impaired fertility

Normal fertility in females but uterine prolapse and fetal malpresentation

2. Epispadias

Less common cf. bladder exstrophy (1:120,000)

Boys > girls 5:1

May be glanular or penopubic

Features

Open urethral plate

Dorsal chordee and lack of dorsal foreskin in males
Bifid clitoris (manta ray) and patulous urethra in females
Pubic symphysis diastasis
Incontinence in all females and 70% males (only glanular continent)

Management

Surgical repair at 1-2 years e.g. Cantwell-Ransley

Principles

Penile lengthening

Correction of chordee (inverting caverno-cavernostomy)

Urethroplasty

Skin coverage (pedicled prepuce island flap)

Additional surgical procedures to improve continence in ~70% at 4-5 years

3. Cloacal exstrophy

Rare (1:200,000)

Boys > girls

Combined bilateral hemibladder and central ileocaecal exstrophy

Often vestigial penis and absent testes

Frequent co-existent congenital anomalies

Management

Termination of pregnancy

Immediate management

Cover with saline packs and either clingfilm or a silo

Give prophylactic antibiotics (Trimethoprim 2mg/kg)

Refer Leeds or Great Ormond Street

Terminal colostomy and bladder repair as for classic exstrophy

Male to female gender assignment (controversial)

Anorectal malformations

1:5000

Males > females 3:2

May be high (above levator) or low (below levator)

High fistulas always associated with urinary fistula, low fistulas sometimes not

Two thirds of boys have high fistulas; two-thirds of girls low fistulas

Typically boys have worse functional outcome (except girls with cloacal malformation)

Commonly a/w other congenital abnormalities, often as a syndrome

Commonest syndrome VACTERL

V – vertebral* (sacral agenesis)

A – anorectal*

C – cardiac

TE – tracheo-oesophageal

R – renal* (VUR)

L – Limb (typically radial)

* commonest

Investigation of newborn with anorectal abnormality

V - screen by plain x-ray spine and spinal USS

R – renal tract USS/MCUG/MAG3

Management of anorectal abnormalities (many surgical procedures)

e.g. Sigmoid colostomy + posterior sagittal anorectoplasty (Pena)

CHARGE syndrome (Coloboma, heart, choanal atresia, mental retardation, genital, ear)